Common Cloaca with Prune Belly like Syndrome

Kedar Ganla, Ajay Mehta, Safala Shroff, S.V. Desai, D. S. Shah, K. M. Ingle
N. W. Maternity Hospital, Mumbai 400 012.

A 22 year old primigravida with 29 weeks gestation was admitted for investigations of Intra uterine growth retardation. There was no prior history of congenital malformation in self or family. There was no history of teratogenic insult.

Her general condition was unremarkable. Obstetric examination revealed uterine size corresponding to 24 weeks of gestation, strongly suggestive of fetal growth retardation. Ultrasound revealed a single live fetus with gestational age of 25 weeks with severe oligohydramnios (AFI-3 cms).

The fetus had an anechoic cystic mass of 12x7 cms, completely filling the lower abdominal region with plenty of echogenic particles within the cystic mass. The bladder was not visualised separately. There were distended bowel shadows seen outside the cystic mass. Other fetal anatomy could not be clealy visualised due to oligohydramnios.

On the basis of ultrasound findings the provisional diagnosis of Meconium Peritonitis with bowel obstruction with the possibility of bladder outlet obstruction was made.

Joint consultation with the Paediatric surgeon revealed poor prognosis which was explained to the patient and the pregnancy terminated after informed consent.

The patient delivered a fresh still born male child weighing 1000 gms. Fetal examination revealed dismorphic facial features, club foot, undescended testis and imperforate anus.

There was gross distention of the fetal abdomen with traumatic rupture of the thinned out anterior abdominal wall which had ruptured during the process of labour, through which the cystic mass had protruded out. Perinatal autopsy revealed a large cystic mass in the lower abdomen. Both the Kidneys were normal and both ureters opened into the side of the cyst on the anterior superior aspect with the urethra opening into it's inferior aspect. The sigmoid colon also opened into the cyst. The cyst contained urine with flakes of meconium in it as shown in the picture.

On the basis of this perinatal autopsy report a final diagnosis of persistent cloaca with Prune belly like syndrome was made.

Persistence of cloaca is an extremely rare malformation occurring in 1:250,000 live births and common in females. It is commonly associated with vertebral and neural tube defects. At about 6 weeks of gestation the cloaca is divided by the urorectal septum into the anterior urogenital sinus and the posterior rectum. Failure of this results in persistence of the cloaca.

The diagnosis of Prune Belly like syndrome was made in view of gross abdominal wall distention thought to be due to bladder outlet obstruction unlike in the true Prune Belly like syndrome where there is significant back pressure changes with gross hydronephrosis of ureters and kidneys. In our case these structures were normal possibly because of good distending capacity of the common cloaca. Our case is an extremely rare congenital malformation and it highlights the importance of perinatal autopsy in arriving at an accurate diagnosis.